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NIH Fiche to Paper Journal

TITLE:	EAR, NOSE, _THROAT JOURNAL
PUBLISHER/PLACE:	Medquests Communications Cleveland Oh
VOLUME/ISSUE/PAGES:	1994 Dec;73(12):926-7 926-7
DATE:	1994
AUTHOR OF ARTICLE:	Isenberg SF
TITLE OF ARTICLE:	Endoscopic approach to the diagnosis of fibrous dy
ISSN:	0145-5613
OTHER NOS/LETTERS:	Library reports holding volume or year 7701817 7882885
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Endoscopic Approach to the Diagnosis of Fibrous Dysplasia

Steven F. Isenberg, MD[a]

Indianapolis, Indiana

A 15-year-old boy had a twelve-day history of severe headaches over the frontal portion of his skull. He had no history of injury or visual changes, and no history of epistaxis. He did have a history of less severe headaches in the past which he had considered sinus headaches. He had been playing in a football game and experienced such a severe headache that he took himself out of the game.

The physical examination was unremarkable with no evidence of cerebrospinal fluid (CSF) leak. Pneumoencephalus was demonstrated on computerized tomography (CT) of the brain (Figure 1). Sinus CT demonstrated an ossified mass in the medial posterior right frontal sinus involving the ethmoid, expansile into the intracranial fossa via the anterior floor of the middle fossa (Figure 2).

An endoscopic approach was designed to biopsy the lesion because it appeared a portion of the mass could be reached by an endoscopic intranasal ethmoidectomy. (Figure 2) Subsequently, the bulla ethmoidalis was entered on the right side and two anterior ethmoid air cells were explored, and thickened mucosa removed. A mass of irregular appearing bone was then encountered hanging from the roof of the ethmoid in the shape of a stalactite. It was subsequently biopsied with cutting forceps under direct visualization. Postoperatively the patient had an uneventful course. The lesion was determined to represent fibrous dysplasia (Figure 3).

Four weeks later the lesion was definitively and completely resected by an intracranial-extracranial approach. A bi-frontal craniotomy resection of the right cribiform plate and planum sphenoidale, along with an external ethmoidectomy and sphenoidotomy, was performed. The patient had one convulsion in the postoperative period but otherwise had an uneventful recovery. Computerized tomography of the brain prior to the definitive surgical approach demonstrated complete resolution of the pneumoencephalus.

[a]St. Vincent and Community Hospitals of Indianapolis, Indiana University Medical Center, Indianapolis, Indiana.

Presented at the Annual Meeting of the American Academy of Otolaryngology -- Head and Neck Surgery, Minneapolis, MN, October 1993.

Reprint requests to: Steven F. Isenberg, MD, 1400 No. Ritter Avenue, Suite #221, Indianapolis, IN 46219-3046.

Discussion

Paranasal sinus masses can be divided into benign and malignant tumors of epithelial or fibro-osseous origin. The benign fibro-osseous lesions can be further subdivided into fibrous dysplasia and ossifying fibroma.

Fibrous dysplasia characteristically manifests itself in childhood and becomes dormant in early adult life. It is of unknown etiology, and is monostotic in 30% and polyostotic

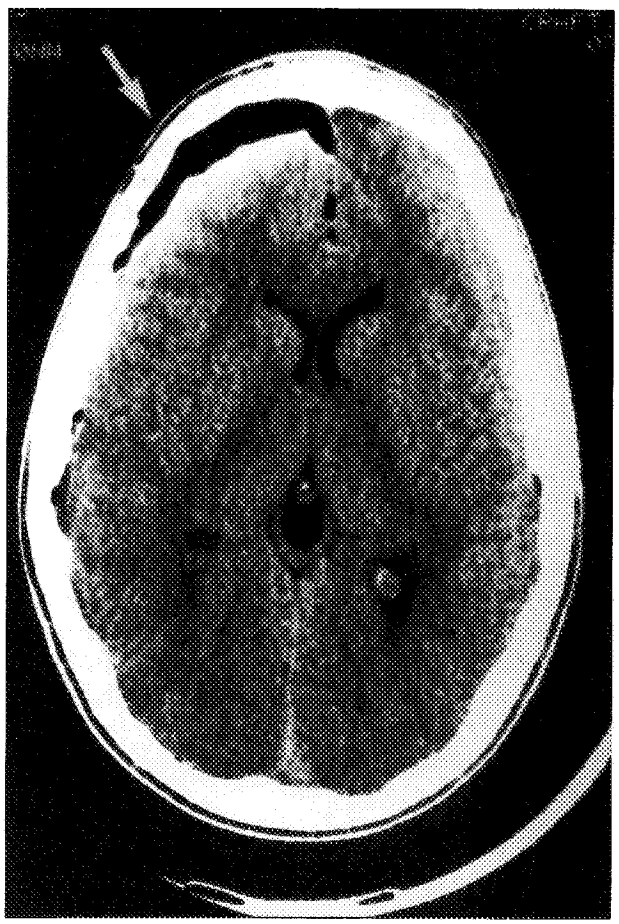

Figure 1. Computerized tomography of the brain. The arrow points to the area of pneumoencephalus.

in 70% of patients. There is no transformation from monostotic to polyostotic form, and malignant transformation usually confined to polyostotic cases, is rare (0.5%).[1] The radiographic appearance looks like ground glass but such an appearance is not diagnostic and biopsy specimens must be taken to differentiate it from similar lesions. Radiotherapy is of no value, and might contribute to malignant transformation.[2] Treatment is by surgical resection or curettage and cosmesis and function should be preserved, if at all possible, since growth of the lesion is usually arrested in early adulthood.[3]

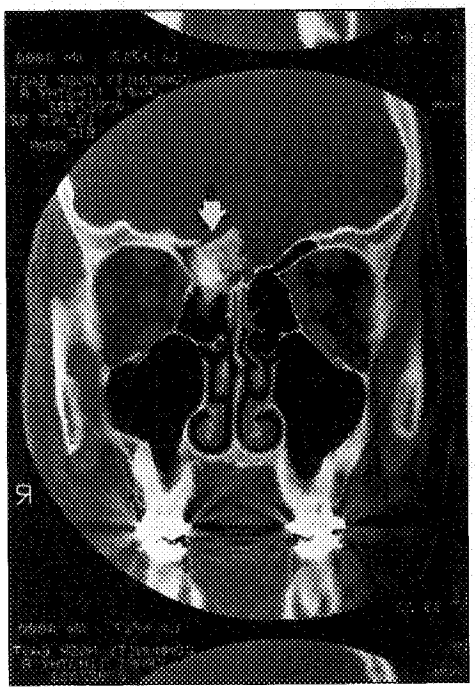

Figure 2. Computerized tomography of the paranasal sinus demonstrating the area of fibrous dysplasia (arrow).

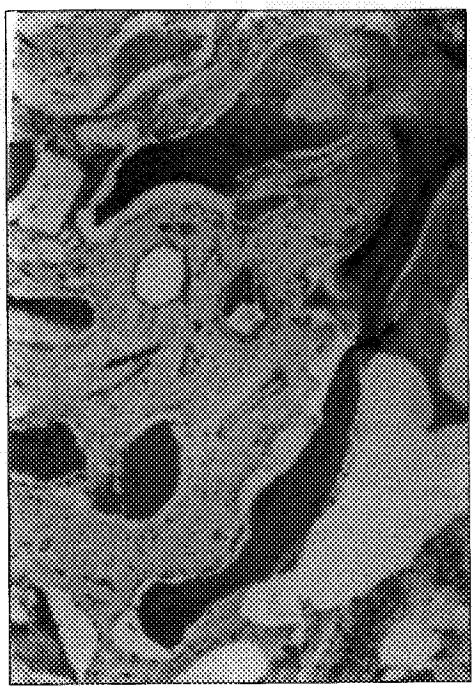

Figure 3. Irregular foci of woven (nonlamellar) bone trabeculae in a cellular but otherwise fibrous stroma. Osteoblastic rimming of bone is minimal. This distinguishes it from ossifying fibroma.

In this case an endoscopic approach was undertaken that successfully led to the diagnosis of fibrous dysplasia without external incisions. Patients with intracranial complications from such a lesion are generally considered candidates for craniofacial resection.[4] The pneumoencephalus in this patient resolved prior to craniofacial resection. Some authors feel that the presence of areas of fibrous dysplasia in the craniofacial bones does not necessarily indicate a need for treatment.[5] Powell[6] reported on fibrous dysplasia of the ethmoid sinus with the diagnosis established by a diagnostic external antrostomy. Both Harrison and Powell noted that surgery in the "active" stage, even to establish the diagnosis, can result in vigorous bleeding with quick regrowth of the lesion. In Powell's case, a definitive surgical resection was performed six weeks after the attempted biospy and the lesion had grown extensively. The attempt at biopsy was non-diagnostic. Harrison[5] states that a deep biopsy obtains more valuable information. An endoscopic approach provides a deep biopsy specimen with minimal tissue trauma, reducing the possibility of stimulating growth of the lesion before a definitive surgical resection or planned clinical observation is undertaken.

An intranasal endoscopic approach to diagnosis allows planning of the appropriate procedure, with the comfort of an established diagnosis. It allows a planned intracranial-extracranial approach in a surgical field that has not been encumbered by previous incisions. Nasal and sinus endoscopes are widely available, since head and neck surgeons are using sinus endoscopes with increasing frequency in the diagnostic and therapeutic management of sinusitis. This report reminds surgeons to consider the use of rigid nasal and sinus endoscopes as a minimally invasive technique for the biopsy of lesions in the paranasal sinuses and anterior cranial base.

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